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(FILE 'HOME' ENTERED AT 17:30:21 ON 09 SEP 2003)

FILE 'MEDLINE, CAPLUS, BIOSIS, SCISEARCH' ENTERED AT 17:30:35 ON 09 SEP 2003

L1 24771 S ELASTIN OR TROPOELASTIN
L2 137016 S TRANSGEN?(6A)(MOUSE OR MICE OR ANIMAL OR COW OR SHEEP OR PIG
L3 59073 S SVAS OR (AORTIC OR CARDIAC)(3A)DISEASE
L4 7 S L1 AND L2 AND L3
L5 4 DUP REM L4 (3 DUPLICATES REMOVED)

=> d bib ab 1-4 l5

L5 ANSWER 1 OF 4 MEDLINE on STN DUPLICATE 1
AN 2003239779 MEDLINE
DN 22625664 PubMed ID: 12626514
TI Domains in **tropoelastin** that mediate **elastin** deposition in vitro and in vivo.
AU Kozel Beth A; Wachi Hiroshi; Davis Elaine C; Mecham Robert P
CS Department of Cell Biology and Physiology, Washington University School of Medicine, St. Louis, Missouri 63110, USA.
NC HL53325 (NHLBI)
HL61006 (NHLBI)
HL62295 (NHLBI)
SO JOURNAL OF BIOLOGICAL CHEMISTRY, (2003 May 16) 278 (20) 18491-8.
Journal code: 2985121R. ISSN: 0021-9258.
CY United States
DT Journal; Article; (JOURNAL ARTICLE)
LA English
FS Priority Journals
EM 200306
ED Entered STN: 20030524
Last Updated on STN: 20030626
Entered Medline: 20030625
AB Elastic fiber assembly is a complicated process involving multiple different proteins and enzyme activities. However, the specific protein-protein interactions that facilitate **elastin** polymerization have not been defined. To identify domains in the **tropoelastin** molecule important for the assembly process, we utilized an in vitro assembly model to map sequences within **tropoelastin** that facilitate its association with fibrillin-containing microfibrils in the extracellular matrix. Our results show that an essential assembly domain is located in the C-terminal region of the molecule, encoded by exons 29-36. Fine mapping studies using an exon deletion strategy and synthetic peptides identified the hydrophobic sequence in exon 30 as a major functional element in this region and suggested that the assembly process is driven by the propensity of this sequence to form beta-sheet structure. **Tropoelastin** molecules lacking the C-terminal assembly domain expressed as **transgenes** in **mice** did not assemble nor did they interfere with assembly of full-length normal mouse **elastin**. In addition to providing important information about **elastin** assembly in general, the results of this study suggest how removal or alteration of the C terminus through stop or frameshift mutations might contribute to the **elastin**-related **diseases** supravalvular **aortic** stenosis and cutis laxa.

L5 ANSWER 2 OF 4 BIOSIS COPYRIGHT 2003 BIOLOGICAL ABSTRACTS INC. on STN
AN 2002:374266 BIOSIS
DN PREV200200374266
TI Dysregulation of TGF-alpha and TGF-beta1 signaling proteins predict clinical features of Costello syndrome.

AU Proud, V. K. (1); Aly, T. A.; Creswick, H. A. (1); Stacey, M. W.
CS (1) Div Med Gen Children's Hosp King's Daughters, Norfolk, VA USA
SO Genetics in Medicine, (May June, 2002) Vol. 4, No. 3, pp. 199.
http://www.geneticsinmedicine.org/. print.
Meeting Info.: Annual Clinical Genetics Meeting of the American College of Medical Genetics New Orleans, Louisiana, USA March 14-17, 2002
ISSN: 1098-3600.
DT Conference
LA English

L5 ANSWER 3 OF 4 MEDLINE on STN
AN 2001316625 MEDLINE
DN 21283166 PubMed ID: 11389427
TI The role of type I collagen in aortic wall strength with a homotrimeric.
AU Vouyouka A G; Pfeiffer B J; Liem T K; Taylor T A; Mudaliar J; Phillips C L
CS Division of Vascular Surgery, and the Department of Biochemistry, University of Missouri, Columbia 65212, USA.
SO JOURNAL OF VASCULAR SURGERY, (2001 Jun) 33 (6) 1263-70.
Journal code: 8407742. ISSN: 0741-5214.
CY United States
DT Journal; Article; (JOURNAL ARTICLE)
LA English
FS Priority Journals
EM 200107
ED Entered STN: 20010709
Last Updated on STN: 20010709
Entered Medline: 20010705
AB PURPOSE: **Elastin** and collagen (types I and III) are the primary load-bearing elements in aortic tissue. Deficiencies and derangements in **elastin** and type III collagen have been associated with the development of aneurysmal disease. However, the role of type I collagen is less well defined. The purpose of this study was to define the role of type I collagen in maintaining biomechanical integrity in the thoracic aorta, with a mouse model that produces homotrimeric type I collagen [alpha1(I)]3, rather than the normally present heterotrimeric [alpha1(I)]2 alpha2(I) type I collagen isotype. METHODS: Ascending and descending thoracic aortas from homozygous (oim/oim), heterozygous (oim /+), and wildtype (+/+) mice were harvested. Circumferential and longitudinal load-extension curves were used as a means of determining maximum breaking strength (Fmax) and incremental elastic modulus (IEM). Histologic analyses and hydroxyproline assays were performed as a means of determining collagen organization and content. RESULTS: Circumferentially, the ascending and descending aortas of oim /oim mice demonstrated significantly reduced Fmax, with an Fmax of only 60% and 23%, respectively, of wildtype mice aortas. Oim/oim descending aortas demonstrated significantly greater compliance (decreased IEM), and the ascending aortas also exhibited a trend toward increased compliance. Reduced breaking strength was also demonstrated with longitudinal extension of the descending aorta. CONCLUSION: The presence of homotrimeric type I collagen isotype (absence of alpha2(I) collagen) significantly weakens the aorta. This study demonstrates the integral role of type I collagen in the biomechanical and functional properties of the aorta and may help to elucidate the role of collagen in the development of aneurysmal **aortic disease** or dissection.

L5 ANSWER 4 OF 4 BIOSIS COPYRIGHT 2003 BIOLOGICAL ABSTRACTS INC. on STN
AN 1994:401736 BIOSIS
DN PREV199497414736
TI **Aortic disease** in **transgenic mice** containing **elastin** gene mutations.
AU Sechler, Jan L.; Boyd, Charles D.
CS AMDNJ-Robert Wood Johnson Med. Sch., New Brunwsick, NJ USA
SO Journal of Vascular Surgery, (1994) Vol. 20, No. 1, pp. 155-156.

ISSN: 0741-5214.
DT Article
LA English

=> FIL STNGUIDE

COST IN U.S. DOLLARS	SINCE FILE ENTRY	TOTAL SESSION
FULL ESTIMATED COST	32.04	32.25

FILE 'STNGUIDE' ENTERED AT 17:33:41 ON 09 SEP 2003

FILE CONTAINS CURRENT INFORMATION.
LAST RELOADED: Sep 5, 2003 (20030905/UP).

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